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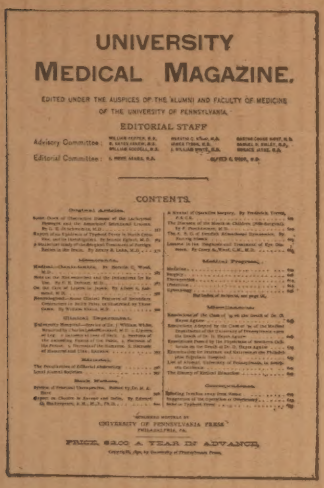

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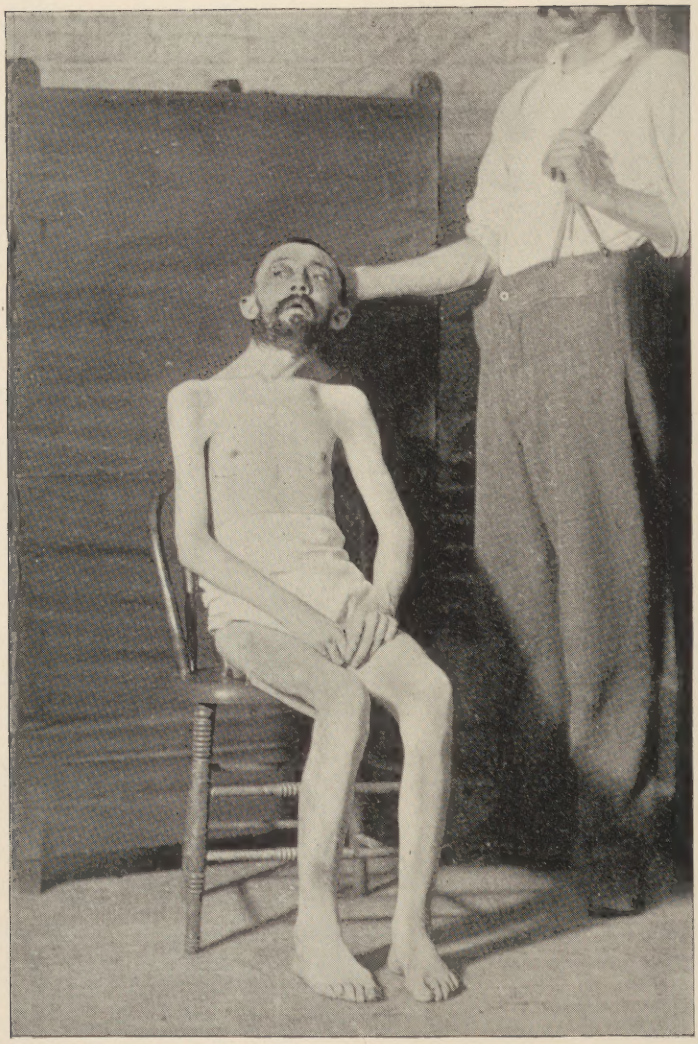
MARCH, 1893

REPORT OF A CASE OF SYRINGOMYELIA, WITH EXHIBITION OF SECTIONS OF THE SPINAL CORD.[1]

By James Hendrie Lloyd, A.M., M.D.

Physician to the Philadelphia Hospital, to the Methodist Episcopal Hospital, and to the Home for Crippled Children.

FIG. 1.



A CASE OF SYRINGOMYELIA.

REPORT OF A CASE OF SYRINGOMYELIA, WITH EXHIBITION OF SECTIONS OF THE SPINAL CORD.[1]

By James Hendrie Lloyd, A.M., M.D.

Physician to the Philadelphia Hospital, to the Methodist Episcopal Hospital, and to the Home for Crippled Children.

I am indebted to the recent monograph of Dr. Bruhl,[2] of Paris, for a brief but clear historical statement of syringomyelia. Ollivier coined the word and applied it in 1837 to all canals or cavities in the cord, the presence of a canal being considered by him as always pathological. Stilling, however, showed that the central canal of the cord is normal and constant. Then the word hydro-myelia was used by Virchow and by Leyden to designate cavities in the cord, which they believed were always expansions of the normal central canal. Hallopeau, in 1869, taught that such cavities were secondary to a myelitis or sclerosis about the natural ependyma. But in spite of theory and imperfect observation, cavities in the spinal cord occasionally showed themselves entirely distinct from the central canal. Then Simon, in 1875, pointed out that cavities result from the softening and breaking down of a gliomatous tissue; that this tissue is a new, degenerative formation, which may exist even as a glioma, teleangiectasic in character; and he proposed to reserve the term "syringomyelia" for these cysts formed in gliomatous tissue and independent of the central canal. This distinction is now generally recognized, and syringomyelia is regarded as the product of a true gliomatosis.

Syringomyelia, having been thus recognized anatomically, has now entered fully into its clinical phase. A long series of memoirs, says Bruhl, seek to establish that it has its own proper symptomatology, and that the diagnosis can be made at the bedside. He adds that an autopsy in many cases has verified the exactness of the diagnosis.

These are precisely the conditions that existed in my own case, for the diagnosis was made upon the patient's admission to the hospital,

[1] Read before the College of Physicians of Philadelphia, February 1st, 1893.

[2] Contribution a l'Étude de la Syringomyélie, par le Docteur I. Bruhl, Paris, 1890.

and the autopsy two months later revealed the extensive and interesting cavity in the cord, sections showing which I have the honor to present to you this evening.

I desire first to state briefly the main features of the symptomatology upon which the diagnosis of this affection rests. Following the admirable classification of Charcot,[1] we note two main groups: (1) The *intrinsic* symptoms, or those related to the limited lesion in the central gray substance of the cord. Here we distinguish (*a*) the symptoms of anterior poliomyelitis—progressive muscular atrophy of the type Aran-Duchenne; (*b*) the symptoms of posterior polio-myelitis—anæsthesia to pain and to heat and cold, with preservation of tactile sensibility and of the muscular sense (this is the well-known dissociation symptom); (*c*) symptoms of central poliomyelitis, a group whose origin is very problematical, including diverse trophic disorders other than those referable to the muscular system. (2) The *extrinsic* symptoms, or those which do not result directly from the gliomatosis in the gray matter, but are frequently associated with the others; they result from the extension of the lesion to the white substance of the cord, and are secondary. They include symptoms of lateral sclerosis, as spastic paresis or paralysis, and those of posterior sclerosis, or tabes. While this classification, like most others, is perhaps artificial in some respects, it is helpful to those who for the first time attempt to master the rather complicated symptom-groups of this affection. It strikes me as doubtful whether the symptoms of lateral sclerosis should be regarded as always secondary, and whether the muscular atrophy is always of the type Aran-Duchenne. In my own case the type, especially on one side, was spastic. It may also be questioned, as in fact Charcot does question, whether the trophic lesions are due to the involvement of the mid-region of the gray matter of the cord. It is evident that we must admit varieties or distinct types of the affection, as in fact the very nature of such a variable and destructive process suggests. In the main, however, the diagnosis rests upon the recognition of certain groupings of the above-named elements. The most common grouping is that of muscular atrophy, especially in the shoulders and arms, with the peculiar "dissociation" sensory symptoms, and with a variety of trophic disorders. My case is as follows:

E. B., white, male, aged 31 years, was admitted into the Philadelphia Hospital in July, 1892. His family history was negative. The patient had not used alcohol to excess, and denied syphilis. He said that he had always been strong until the winter of 1888, when after a severe storm he had a swelling of his right ankle, which he

[1] Bruhl, *op. cit.*

regarded as rheumatic. He noticed soon afterward that he was obliged to step on his toes, and that his shoes became worn on the front parts of the soles. He was in the hospital for a few weeks in 1889. His condition on admission last July was as follows:

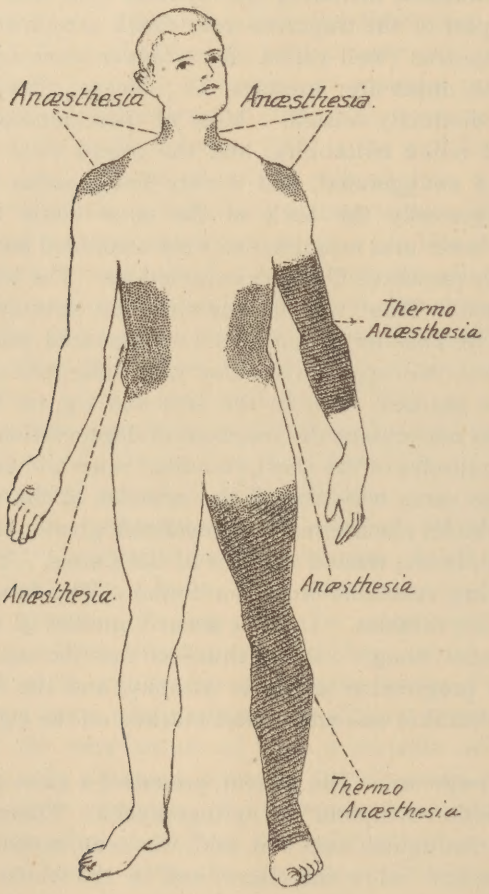
Motor Symptoms.—The patient had advanced progressive muscular atrophy of the spastic type (the so-called amyotrophic lateral sclerosis). The shoulder muscles, including the deltoid, infra- and supra- spinati and the lower part of the trapezius were much atrophied. The upper part of the trapezius (well called the *ultimum moriens*) was in good condition. The upper-arm muscles, *i.e.*, biceps, triceps and deeper muscles, were distinctly wasted. Most of these muscles presented a high degree of reflex irritability, and the biceps were spastic. The biceps jerk was exaggerated, and a very lively reflex was produced by tapping vigorously the back of the wrist while the hand was dropped. The lower-arm muscles also were atrophied and weak. Some of these muscles presented fibrillary contractions. The legs were spastic and contractured. There was no true muscular atrophy or fibrillation in the legs. The knee-jerks were much exaggerated, and ankle clonus was present; but this spastic condition with exaggerated reflexes was distinctly more marked, both in the arm and leg, on the right side. The muscles did not present the reactions of degeneration. To faradism all the wasted muscles of the chest, shoulder, arms and forearms reacted normally. The same was true of the muscles of the legs. To galvanism the cathodal closure reaction remained greater than the anodal closure reaction in the wasted muscles of both arms. The anodal and cathodal opening reactions were not found. The formula remained normal in the leg muscles. In some wasted muscles of the arms there was a slight modal change. It was thus seen that the case presented the spastic type of progressive muscular atrophy, and the interesting fact was observed that this was much more marked on the right than on the left side.

Sensory Symptoms.—The patient presented a quite typical picture of the "dissociation-symptom" of syringomyelia. Thermo-anæsthesia, or inability to distinguish heat and cold, was seen, especially in the left arm both above and below the elbow, and in the whole of the left leg both anteriorly and posteriorly; also upon the back (Figs. 2 and 3). Analgesia, or loss of sensibility to pain, occupied almost the same areas excepting the back (Figs. 4 and 5). Small areas of anæsthesia on the neck, shoulders and front of the waist were also seen. (See Figures.) This anæsthesia is not common in the disease, and in this case occupied but a limited area. On the right thigh, instead of analgesia there was hyperæsthesia. This observation is perhaps of some significance in view of the fact that the sensory symptoms peculiar to

syringomyelia were more marked on the left side, while the motor symptoms were more marked on the right. These sensory symptoms show the segmental distribution commonly seen in this disease.

Deviation of the spine was a noticeable feature in this patient. It is a very common symptom of syringomyelia and, according to Bruhl,

FIG. 2.



SYRINGOMYELIA.

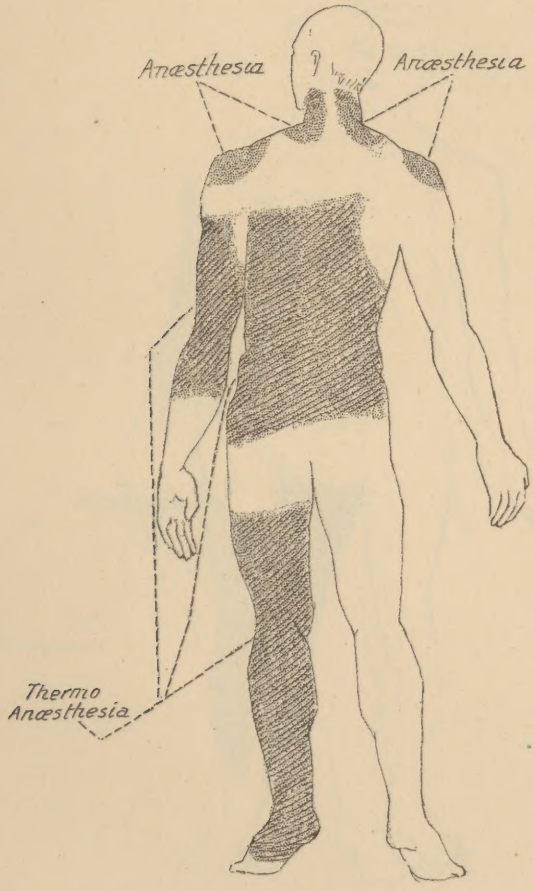
Showing Segmental Distribution of Sensory Symptoms.

exists in at least 50 per cent. of cases. This scoliosis, in my case, affected even the cervical spine and the position of the head, giving the neck and head a twist not unlike that seen in torticollis. This is shown to advantage in the photograph (Fig. 1), which shows also the weakness of the neck muscles, which made it necessary for the patient to have his

head supported by an attendant while being photographed. When the patient was lying down, this scoliosis was almost obliterated.

Trophic Disorders.—The right ankle was enlarged, but not so much so as formerly, according to the patient. It was undoubtedly an arthropathy similar to those seen in locomotor ataxia, general paresis and multiple sclerosis, to which diseases and to syringomyelia true spinal

FIG. 3.



SYRINGOMYELIA.

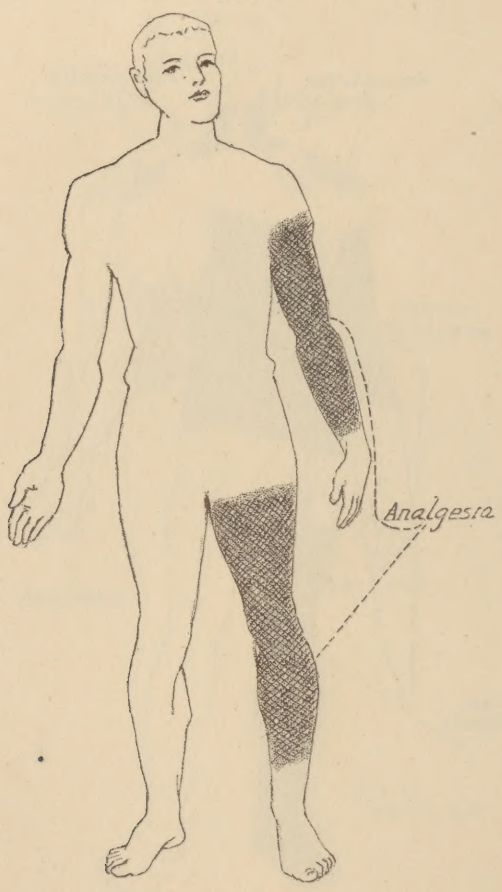
Showing Segmental Distribution of Sensory Symptoms.

arthropathy appears to be limited. Grating, denudation of bone, stalactites and effusion were not noted. It is probable that the arthropathy had been of the milder type, recognized by many original observers, in which there is a tendency to recover. This enlarged ankle is seen in the photograph.

The toe-nails were very much deformed; they were enlarged and thickened, had transverse ridges and were quite brittle. The patient said that several times some of his nails had dropped off.

The man had deeply pigmented maculæ on the legs, which can be distinguished in the photograph. These spots and blemishes were the results of a slowly advancing trophic process in the skin. These

FIG. 4.



SYRINGOMYELIA.

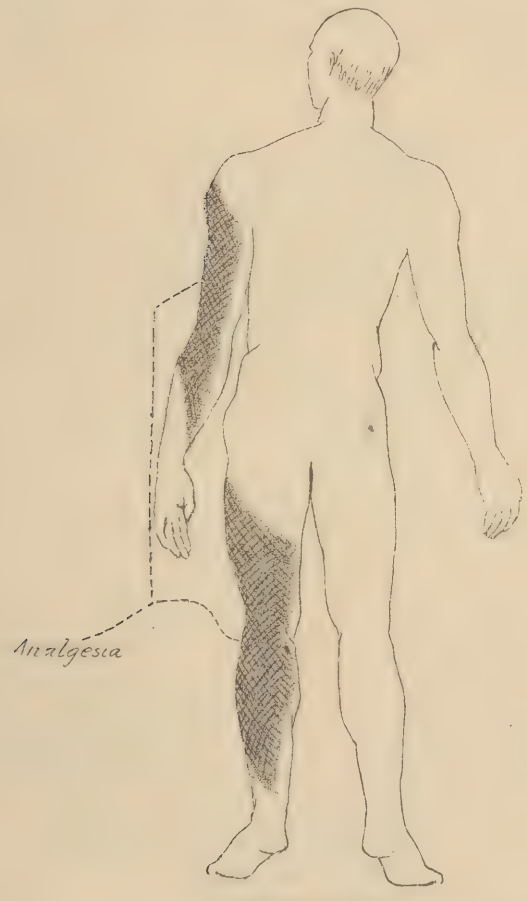
Showing Segmental Distribution of Sensory Symptoms.

changes in the nails and skin have been observed in other cases of syringomyelia. Like most trophic disorders dependent upon central nervous disease, they do not appear to follow any constant law, and their exact pathogeny is obscure. In Morvan's disease, however, which is now generally recognized as only a special type of syringo-

myelia, trophic changes, leading to total destruction of the fingers, the painless whitlow or "panaris analgesique," of French writers, are characteristic.

No mental symptoms were observed, nor any symptoms of bulbar involvement. Expansion of the chest was limited to not quite one inch, the left side of the chest being rather freer than the right. I have

FIG. 5.



SYRINGOMYELIA.

Showing Segmental Distribution of Sensory Symptoms.

made a similar observation several times in multiple neuritis, but never before in a case of cord disease. There were no oculo-motor symptoms. The bladder and bowels were innervated normally. The heart was rapid, the sounds normal.

Soon after admission the patient's temperature was noted occa-

sionally to be increased—sometimes to the height of 102°. The lungs were carefully examined, but no evidence of phthisis was found. During August he had an attack of dysentery—which was prevalent at that time in the hospital. This weakened him so much that he was not able afterward to leave his bed even to be propped in a chair. In September I found on visiting him one day that he had a severe cough which had begun on the preceding day. Examination revealed pneumonia, under which he sank rapidly and died the same evening. His embarrassed respiration was doubtless the cause of the rapid progress of the lung complication.

At the autopsy, the cord presented the characteristic appearance described by Bruhl. The cervical enlargement was broadened and flattened (somewhat ribbon-like), and to the finger, gently pressing or squeezing it, gave the impression of containing a cavity. On section a large cavity was found beginning in the very lower part of the medulla, broadening out in the cervical region, and extending well down into the dorsal cord. It did not extend into the lumbar enlargement. This canal, or cavity, was very wide in the cervical enlargement; so wide in the fresh state that it is not an exaggeration to say that a small penholder could have been inserted into it. In this region it had caused great destruction or alteration of arrangement of the normal constituents of the cord, as seen even by the naked eye. This cavity was not exactly in the middle, but extended rather more to the right side—a fact which corresponds to the clinical findings. There were no special signs of inflammation, or of hemorrhage, about the cord.

Sections of the cord have been made from fourteen levels. For this work I am indebted to Dr. Albert A. Ghriskey and Dr. James Homer Wright, of the laboratory of hygiene in the University of Pennsylvania.

I wish here to particularly acknowledge my obligation for this work, and to call special attention to these beautiful sections, which have been stained by the Weigert method. The photographs of the sections were made by Dr. W. N. Gray, of the U. S. Army Medical Museum in Washington, to whom also I am much indebted. The drawings (Figs. 6, 7 and 11) were made by Dr. A. A. Stevens.

DESCRIPTION OF SECTIONS.[1]

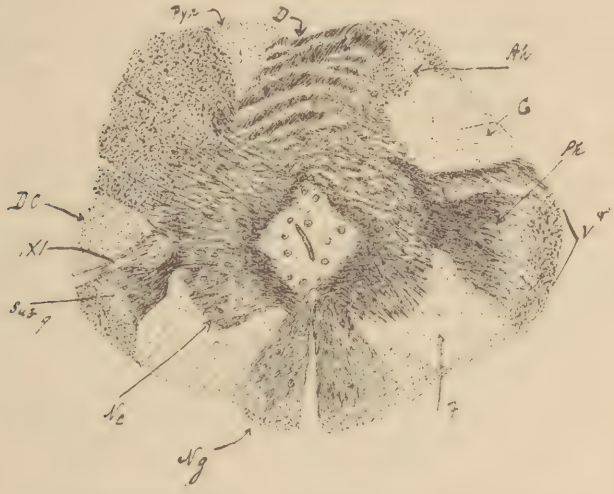
General Features.

Under a high power the glioma, stained with methylene blue, presents numerous neurogliar cells. With Weigert's stain these are not quite so readily seen. They are well seen with a carmine stain. The fibrillary structure, described by some authors, does not appear plainly in these sections. It is apparent in some limited spots, where the tissue is not so dense as usual; but I have not been able to follow fibrillæ from their cells. Nevertheless, the glioma has a mesh-like formation,

[1] The author is entirely responsible for these descriptions, to which he has given much attention and care, especially to that of the medulla oblongata, in which the anatomy is complicated and still further confused by disease.

which shows innumerable minute spaces. Very few vessels are seen in the new formation, and nothing like the single longitudinal vessel of supply described by Bäumler. Most of the vessels are at or near the periphery of the mass, but at least one well-defined one is seen in the cavity. Refractive bodies, described by Bruhl, are plainly visible about and beyond the periphery of the new formation. The cavity has, at some points, what seems to be a lining membrane, which is even detached in some places, as described by others. This appearance is evidently caused by the shrinking away from the cavity-wall of the solid transparent contents of the cavity, carrying with it some adherent portions of the glia. At some levels a large group of broken-down epithelial cells, evidently the remains of the central canal, can be seen. This is situated in front of the cavity, and to one side

FIG. 6.



SYRINGOMYELIA. (*MEDULLA OBLONGATA*).

Ng.	Nucleus funiculi gracilis.	*V.*	Ascending root of fifth nerve.
Nc.	Nucleus funiculi cuniati.	*Ph.*	Posterior horn.
F.	Funiculus cuniatus.	*G.*	Gower's tract.
Sub. g.	Substantia gelatinosa.	*Ah.*	Anterior horn.
XI.	Accessory nerve.	*D.*	Decussation of pyramids.
DC.	Direct cerebellar tract.	*Pyr.*	Left pyramid.

The light, unshaded areas are degenerated. The central canal is surrounded with gliomatous material. The fibres of the accessory nerve are too diagrammatic. They are not so distinct in the section.

of the centre. It is not connected with the cavity. In the medulla, however, and again in the lumbar enlargement, the central canal, surrounded by gliomatous new formation, can be made out more plainly. There is but little pigment in the cord anywhere. No conspicuous signs of inflammation or hemorrhage can be seen; the membranes are not unduly thickened, nor are the bloodvessels enlarged, increased nor crowded with leucocytes. Most of the multipolar cells in the anterior horns of the cervical enlargement are atrophied and granular. The lateral pyramidal tracts are densely degenerated, and contain only a few scattered nerve fibres in varying stages of decay. The direct cerebellar tracts, to the outside of the crossed pyramidal tracts, are much better preserved than the latter. At some levels they

contain a sufficient number of normal fibres to make a contrast to the adjacent pyramidal tracts visible in the mounted section to the naked eye. This is significant in view of the connection of these fibres with Clark's vesicular column, and their possible influence on equilibration—which was not disturbed in this patient. They are not so well preserved on the right as on the left side; and in the medulla at the level of the decussation of the pyramids, the region of the direct cerebellar tract, as well as of the pyramidal tract before it decussates, is degenerated. The integrity of Clark's vesicular column is difficult to determine, because the region in which it is found is stretched around the end of the cavity in such a way as to distort and confuse the nerve elements.

This cord, followed through its whole length, presents an epitome of the gliomatous process in all its various stages. Thus, in the medulla the process is diffused in various areas, and a cavity has not yet been formed. In the cervical region the cavity is formed, and is very extensive, with secondary effects in the white matter; in the dorsal region the process is more limited, and the glioma tends to one side; while in the lumbar enlargement the process is still in an early stage, prior to the formation of a cavity.

SECTION 1 (Fig. 6).—This is the highest section made—about the region of the decussation of the pyramids. The central canal, lined with epithelium, is seen. It is surrounded by gliomatous material, which has not yet begun to break down into a cavity, although under the microscope it is seen to be brittle and friable. The decussating fibres are seen and the remnants of the anterior horns cut off by them, lying to their outer side. It is to be noted that the fibres running toward the right are much more numerous than those running in the opposite direction; also, that the left pyramid is degenerated.[1] The nuclei of the funiculus gracilis and funiculus cuniatus are well preserved. The appearance of cells and ganglion formation can be made out in both. The funiculus cuniatus itself, lying at the head of the column of Burdach, is deeply degenerated on each side. The funiculus gracilis, or head of the column of Goll, is much better preserved. The beginning of the reticular process is visible, although much broken up by glia. The substantia gelatinosa can be seen around the ends of the posterior horns. These posterior horns are of different size and appearance, the left being much the smaller. To either side of the central region (here occupied by glia) is a collection of gray matter with some large cells; and from this region fibres arise and run toward the side of the cord, skirting along the edge of substantia gelatinosa. These are doubtless the fibres of the accessory nerve. In the right lateral region of the cord, occupied by both the direct cerebellar tract and Gower's tract, the cord is much more degenerated than in the corresponding region on the left. On the opposite side of the cord a small degenerated area is seen also in front of the posterior horn. This is the left direct cerebellar tract, showing here to advantage its triangular shape.

Just below this region a section (not shown in the illustrations) shows extensive gliomatosis, with a very large cavity. The new formation occupies not only the central region but also portions of the right lateral column and of the posterior columns. The left antero-lateral column is partially preserved. But little gray matter is visible, the cord being a hollow tube with some conducting tracts of white matter preserved.

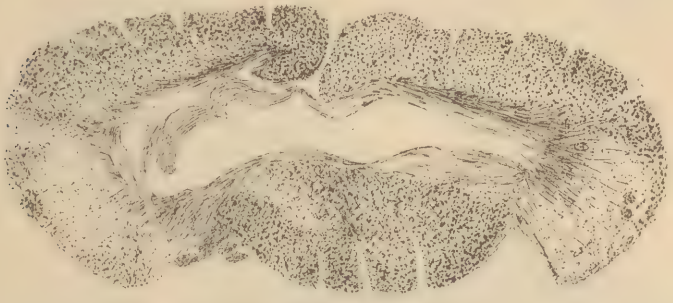
SECTION 3 (Fig. 7).—Upper cervical region. Here the cavity extends far across the cord, almost symmetrically on either side. The gliomatous formation extends out along the posterior horns, almost cutting off the posterior columns from the rest of the cord. The lateral tracts are much degenerated, especially the right,

[1] This degeneration of the left pyramid *above* the cavity is worthy of special note. It will be remembered that the patient's motor symptoms were more marked on the right side.

and the right direct pyramidal tract. Remnants of the anterior horns can be made out as narrow projections in front of the cavity (the left alone is shown in the drawing); they contain only a few multipolar cells degenerated. The anterior parts, especially, of the posterior columns, are degenerated. But little trace of posterior root-zone. The gliomatous tissue extends almost entirely around the cavity as a rim or circle.

[NOTE.—Many of these general features are characteristic of the segments lower down. The cavity tends to run to the right side in the dorsal cord (as we shall see) and to diminish in size. The lateral tracts will be observed in all the sections to be degenerated to the very end of the cord. The anterior horns are degenerated only in the cervical region. They appear with a full complement of cells in the lumbar enlargement. From the region of the decussation, first described, the central canal is partly obliterated; its epithelial lining, however, is found in almost every section as a group of epithelial cells not connected with the cavity. The cord is distinctly smaller in the cervical region on the right than on the left side. In all, fourteen sections from different levels of the cord were mounted and have been carefully examined. In order, however, to avoid repetition, only a selected number of these are illustrated and described.]

FIG. 7.



SYRINGOMYELIA.

SECTION 4 (Fig. 8).—*Photomicrograph.*—The cavity here begins to trend toward the right side (left in the photograph). It is lined with a gliomatous material. Remains of the central canal can be seen in a group of epithelial cells in front of the cavity and to one side of its normal position. It presents irregularly the outlines of a double canal. It does not appear in the photograph. The anterior white commissure is preserved. The gray matter is stretched around the ends of the cavity and only a few multipolar cells are visible (very indistinct in this photograph) in the anterior horns. The lateral pyramidal tract is deeply degenerated, and the direct pyramidal tract slightly so. The posterior horns and root-zones cannot be well distinguished.

SECTION 5 (Fig. 9.)—*Photomicrograph.*—Cervical region next below Section 4. Cavity trends still further to right. (In this the image is not reversed.) The anterior horn can be well distinguished. It contains more multipolar cells than the former section, but under a high power they are seen to be degenerated. Otherwise the appearances are much the same as in Section 4. The direct pyramidal track on the right is more degenerated than on the left. The anterior portion, especially, of the columns of Goll are involved in gliomatosis.

[NOTE —In the next two segments, not shown here, the cavity again becomes almost symmetrical, before deviating finally in the dorsal region.]

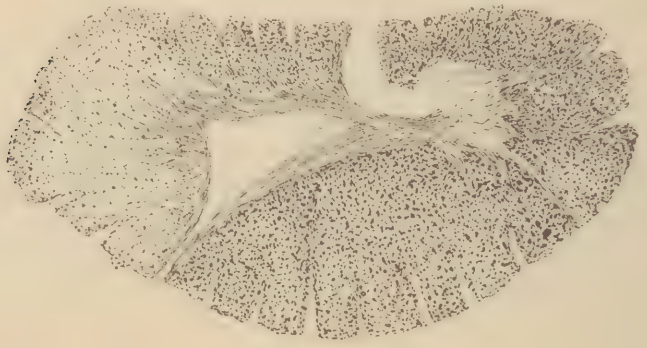
SECTION 9 (Fig. 10.)—*Photomicrograph.*—Upper dorsal. Here the cavity trends

to the right (image not reversed). The gliomatous tissue extends down both posterior horns. All around it the white, as well as gray, matter is degenerated, especially in the direct and crossed pyramidal tracts (more marked in the right). The anterior parts of the posterior columns also are affected. The gray matter is almost entirely destroyed, except the anterior horns, which are seen as mere small projections in front of the cavity. They contain but few cells.

SECTION 12 (Fig. 11).—This drawing represents very well a section from the mid-dorsal region. (The image is reversed.) The cavity is to the right (left in figure), and follows out the posterior horn quite to the periphery. The right lateral tract is very much degenerated, and the left is rather more so than is shown in the drawing. The right anterior horn is also practically destroyed. The left anterior horn preserves its shape, and under the microscope some motor cells are visible in it. The posterior columns show degenerated fibres scattered through them.

SECTION 14 (FIG. 12).—*Photomicrograph.*—From lumbar enlargement. The cord here presents a striking contrast to its appearance higher up. Its general shape

FIG. 11.



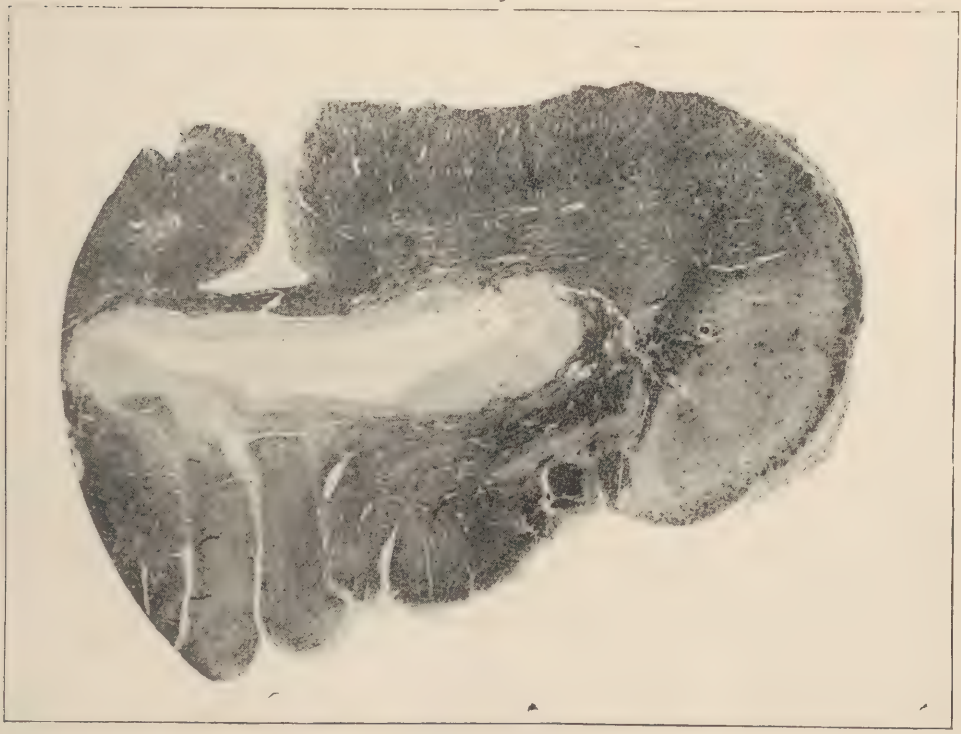
SYRINGOMYELIA.

and size, as well as those of its gray matter, are normal. The anterior horns are large and well supplied with motor cells. The posterior horns and root-zones are plainly seen. In but two features does it present very marked abnormal appearances; and these, in view of the diseased state of the cord higher up, are full of interest. First, the lateral, or crossed, pyramidal tracts, which are here small and have come to the periphery, are degenerated. Second, the region of the central canal, in the posterior gray commissure, shows distinctly gliomatous change. Already the central canal is obliterated, but no gliomatous cavity has yet taken the place of the normal one. This proves very beautifully that the cavity in syringomyelia is *not* a mere expansion of the normal canal. On the contrary, the normal canal does not exist any longer, and its place is taken by the remains of its columnar epithelial cells, which have undergone coagulation necrosis with loss of their characteristic columnar appearance. This group of broken-down cells is surrounded by the gliomatous mass, which at this level occupies but a small space in the gray commissure. We see here, probably, an early stage of the gliomatous process at the lowest point of its downward progression in the cord. Slight degeneration is seen here in the posterior columns near the periphery.

FIG. 8.

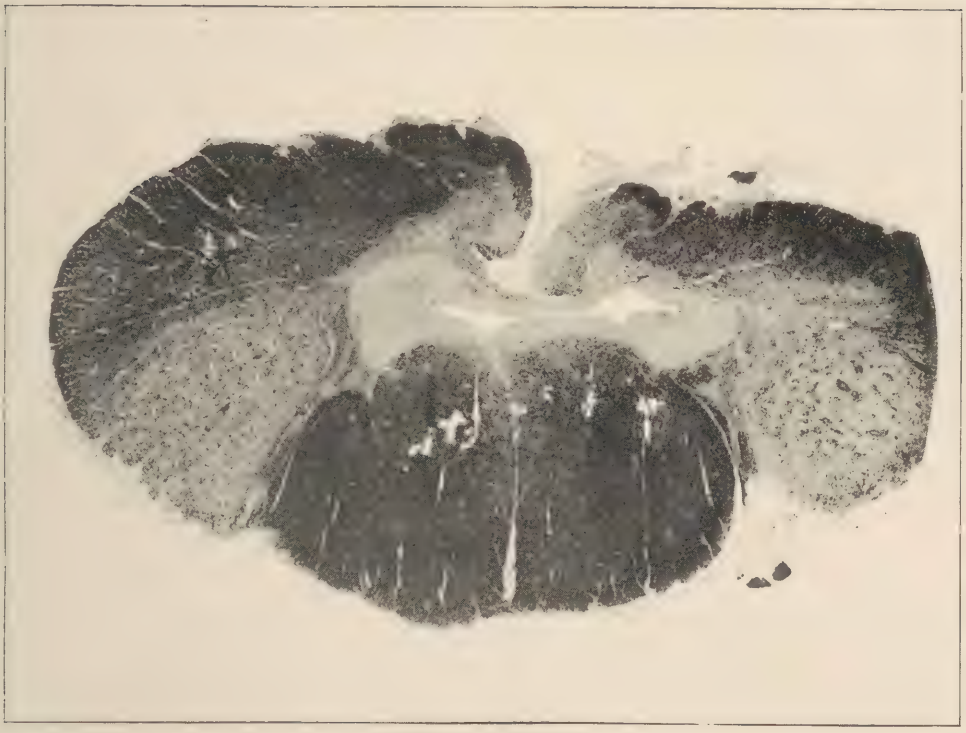
SYRINGOMYELIA.
Cervical enlargement (Section 4).

FIG. 9.



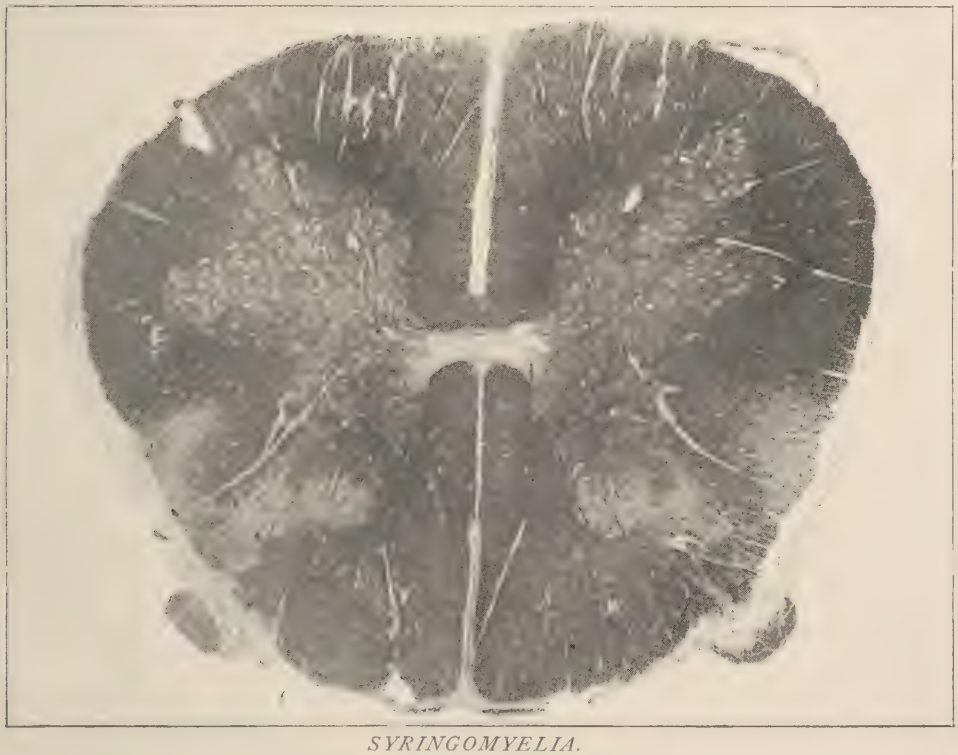
SYRINGOMYELIA.
Cervical enlargement (Section 5).

FIG. 10.



SYRINGOMYELIA.
Upper dorsal region (Section 9).

FIG. 12.



SYRINGOMYELIA.
Lumbar enlargement (Section 14).

A few special points occur to me before closing this report.

Already special types and varieties of syringomyelia begin to appear and to be recognized. These are the "formes frustes" of French writers. The most notable of these is the "type Morvan," which, until very recently, was claimed by many to be a distinct disease. Its special symptom is a destructive whitlow, painless and chronic, of the fingers. Joffroy has already proved that this destructive lesion is trophic, and that it depends upon a gliomatous change leading to syringomyelia. Other trophic lesions sometimes dominate the scene, and by appearing early and constituting the most conspicuous feature, may be confusing in the diagnosis. Joint changes may thus appear, and be called rheumatic. In my own case it may be recalled that an enlargement of the ankle was the first symptom noted by the patient. This was evidently a true spinal arthropathy. Another lesion is the one noted by Charcot of enlargement of the hand, closely simulating that of acromegalia, which Charcot calls chiromegalia. In his case the change was limited to one hand, and was proved to be symptomatic of syringomyelia.

Other varieties are the asymmetrical cases. These may be monoplegic or hemiplegic in type. A case has already been reported in which a hemiplegia of spinal origin was the earliest symptom. This doubtless depends upon the cavity extending, as in my own case, more toward one side.

The possible confusion of syringomyelia and some of the cases of so-called Friedreich's ataxia, has always been an interesting point to me. In some cases of syringomyelia the posterior root-zones and columns are involved (the secondary extrinsic symptoms already referred to). This fact, together with the existence of scoliosis, and the well-recognized fact that both diseases are probably developmental or embryonal in origin, and the still more significant fact that in three out of the twelve autopsies in cases of Friedreich's ataxia, collected by Griffith, cavities were found—all these facts suggest an analogy and a possibly deep relationship between the two diseases. Dejerine contends that Friedreich's ataxia is due to a gliomatous change.

Probably the most widely received opinion of the exact nature of syringomyelia is that it depends upon the proliferation of an embryonal tissue remaining in that region of the cord in which the medullary folds in the embryo close over to form the central canal. It is thus a developmental defect, and the causes of it reach far back into the intrauterine life.

That this degeneration is not a mere sclerosis of connective tissue finds a curious confirmation in a work of Chaslin,[1] who observed some special features in the histo-chemical reaction of this gliomatous material which serve to differentiate it.

[1] Quoted by Bruhl (p. 89).